Sickle Cell Anemia (Signs and Complications)

Many of the signs and complications of sickle cell anemia are due to sickling within microvascular beds leading to vascular occlusion and hypoxia. Individuals with sickle cell disease experience many different crises and conditions. Distinguish aplastic crisis, splenic-sequestration crisis and hyper-hemolytic syndrome from each other based upon whether there is appropriate reticulocytosis or not and the degree of splenic involvement. Auto-splenectomy is the end result of episodes of splenic-sequestration crises and painful crises occur in various parts of the body.



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Signs

Crew-cut on Skull X-ray
Crew-cut Skull
Anemia and subsequent lack of oxygenation results in increased erythropoetic drive leading to marrow hyperplasia. On x-ray, marrow hyperplasia in the flat bones of the skull resembles a crew-cut haircut.

Complications

Auto-splenectomy
Spleen-committing-seppuku
The microvascular beds of the spleen have a slow transit time allowing aggregation of deoxygenated HbS to occur. This results in RBC sickling within splenic vascular beds thus preventing adequate blood flow to the spleen. Over time, hypoxic damage, thrombosis, infarction and fibrosis occur to the spleen causing it to be shrunken and nonfunctional by the age of 3-4 years old in an individual with sickle cell disease.

Painful Crisis
Pain-bolt Crying
These are vaso-occlusive crises due to RBC sickling occurring in a wide variety of locations in the body. These occur in an episodic nature and can be precipitated by infection, dehydration, acidosis and usually unknown causes. Dactylitis is due to vessel occlusion in the hands of the bone; acute chest syndrome (fever, cough, chest pain, pulmonary infiltrates) occurs with lung vessel involvement; stroke can occur with brain vessel involvement and priapism can occur with penile vessel involvement.

Aplastic Crisis
A-plastic-bottle Crying
This is an emergency that occurs when an individual with sickle cell disease becomes infected with Parvovirus B19, which infects erythroid precursors. It results in a severe drop in hemoglobin and reticulocytes to <1%. Unlike aplastic anemia, there is no pancytopenia in aplastic crisis. Normal individuals do not experience aplastic crisis when infected with Parvovirus B19 because they have adequate RBC stores and can tolerate shut-downs in RBC production for 1-2 weeks.

Splenic-sequestration Crisis
Clogged-spleen Crying
This is an emergency due to Intra-splenic trapping of RBCs leading to a drop in hemoglobin with appropriate reticulocytosis and painful enlargement of the spleen. This can result in hypovolemic shock.

Hyper-hemolytic Syndrome
Hiker with Hemolysing RBCs
This is an emergency where there is a drop in hemoglobin with appropriate reticulocytosis and no painful enlargement of the spleen. It occurs due to accelerated RBCs breakdown.

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Renal Papillary Necrosis

Kidney Paper Necrosis-crow

Vaso-occlusion in the renal papillae results in its necrosis and sloughing off. Patients experience acute colicky flank pain, gross hematuria and passage of tissue fragments in urine.

Salmonella osteomyelitis

Salmon Skeleton-in-flames

Due to splenic dysfunction and subsequent auto-splenectomy, patients have increased susceptibility to encapsulated bacteria. Studies have demonstrated that patients suffering from sickle cell disease are more likely to have osteomyelitis due to the encapsulated bacteria Salmonella than other common causes such as S. Aureus.

Pulmonary Hypertension

Lungs Hiker-BP

Vaso-occlusion of pulmonary vessels results in hypoxia and subsequent vasoconstriction (hypoxic pulmonary vasoconstriction) in order to redirect blood flow to alveoli with higher oxygen content.

Hyposthenuria

Hippo-pasta-urinal

It is thought that sickling in the vasa recta of the kidneys impairs countercurrent exchange and free water reabsorption resulting in excessive urination.

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